One glasses too many: A case report of Benson's syndrome

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We report a case of Benson's Syndrome, a form of occipital Alzheimer's disease, with posterior cortical atrophy on magnetic resonance imaging, in a 62-year-old male, who presented with visual problems, ascribed to the eyes, and had even undergone cataract/intraocular lens surgery in the right eye; and change of glasses 21 times over the past 2 years, with no apparent benefit. This case is of interest both on account of its rarity, and to highlight its features since the diagnosis may be missed in an ophthalmological setting where such patient may go for first consult.

Key words: Benson's syndrome, frequent change in glasses, posterior cortical atrophy

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Alzheimer's disease (AD) is an acquired, incurable and progressive disorder that leads to social and occupational malfunctioning on account of impaired cognition and behavior with memory loss as a predominant feature. Pathologically, it is associated with plaques in the cerebral cortex. The criteria for diagnosing AD require the finding of slowly progressive memory loss of insidious onset in a conscious patient and excluding the possibilities of toxic metabolic conditions and cerebral neoplasms. AD can be classified into three stages: Asymptomatic preclinical, mild cognitive impairment, which is a symptomatic predementia phase, and frank AD dementia.[1] A visual variant of AD, Benson's syndrome, also called posterior cortical atrophy is a localized neurodegenerative condition primarily affecting the occipital, parietal and occipito-temporal cortices. First described by Benson *et al*. in 1988,[2] Benson's syndrome is a clinicoradiologic syndrome associated with a decrease in visuospatial and visuoperceptual capabilities with relatively intact language, learning and cognition in the

early stages. Its exact prevalence and incidence are unknown.[3] Despite efforts to develop diagnostic criteria, no clear consensus has emerged. Its co-features include in addition, evidence of complex visual disorders including features of Balint's syndrome (simultanagnosia, saccadic initiation failure, deficit in reaching to visual goals, environmental agnosia) and Gerstmann's syndrome (acalculia, agraphia, finger agnosia, left/right disorientation), field defects and visual agnosia, with an absence of stroke or tumor. Supportive features include a presenile onset (mid-50s or early 60s), alexia, dressing apraxia, prosopagnosia and prolonged color after-images.[4] With an onset much earlier than that for a typical AD, patients usually present to an ophthalmologist with change in vision, difficulty reading and writing, diplopia and problems with depth perception. They have trouble reaching out to pick objects of interest, poor contrast sensitivity and difficulty recognizing colors. Visual field may show hemianopia. Visual crowding or visual hallucinations may also be present. Anterograde memory, linguistic skills and executive functions are preserved in the early stages, but as the disease progresses patients may develop misrecognition of familiar faces and objects, and calculation skills and co-ordinated movements may get affected. Later problems with memory and cognitive impairment may occur. Neuropsychological evaluation, blood tests, brain scans and neurological examination is required to exclude any treatable cause including infection, inflammation or tumor.[5] We report the case of a 62-year-old male diagnosed with Benson's syndrome, of interest due to its extreme rarity,[3] and its largely ophthalmological presentation.

Case Report

A 62-year-old man reported with a history of bilateral progressive, painless blurring of vision for the last 2 years. Two years ago, admittedly for visual problems, he underwent uneventful cataract surgery with intraocular lens implantation in the right eye, but with no visual benefit. He complained of diplopia (binocular) following cataract surgery. He continued to experience difficulty in reading and writing and problems judging distance and depth, as evident in difficulty in reaching out precisely for objects and during ambulation. He also complained of inability to perceive objects in the right visual field: Would miss food on the right half of the plate and not notice the right half of his reflected face in the mirror. He had met with an accident 2 years ago, while riding his motorcycle, injuring his right arm that he confessed was on account of not perceiving objects to his right: He has stopped driving since. His visual difficulties worsened in dim-light; while he also had increased sensitivity to bright light. He had started wearing glasses for presbyopia around 25 years ago, and would periodically get them altered every couple of years, but in the last 2 years, had changed them 21 times with no improvement in his vision. During this period, he consulted both ophthalmologists and optometrists, more often the latter in "optical shops," and was often advised a change of a minor nature, although more often than not, he did not perceive any worthwhile improvement in vision. During this period, his vision continued to deteriorate from a recorded 20/30 Snellen at the time of cataract surgery to the present state. He denied any hallucinations and weakness. There was no family history of dementia, and he was not a hypertensive or diabetic.

On examination, his vitals were stable, and systemic examination was within normal limit. His corrected VA was 20/60 in each eye: With +0.5–2.0 × 90 OD and +1.0–2.0 × 90 OS. The right eye was pseudophakic. Biomicroscopy and ophthalmoscopy were unremarkable; with no RAPD, and a normal fundus [Fig. 1].

He tended to have a head turn to the right, keeping his eyes straight, and appeared to look askance, and clarified that this allowed him to get his better seeing left field on his object of interest. Ocular movements were full and free. There was no misalignment, with symmetrical Bruckner's reflexes. The confrontation tests suggested a right hemianopia. The full-field perimetry revealed a bilateral field defect, denser in the right fields, but involving the periphery of the left fields too [Fig. 2].

This co-relates well with the existing magnetic resonance imaging (August 2014) findings of bilateral occipital cortical atrophy, more marked on the left [Fig. 3].

On Ishihara color plates test, in bright day-light, with the near correction, the patient could barely decipher digit '1' from the '12' available on the explanatory first plate: He failed to notice any other number. On attempting the Pelli-Robson contrast sensitivity function, with great difficulty he could make out the letter-triad with logarithmic contrast sensitivity value of 1.35 each eye. General neurological examination showed no cognitive deficits, no ataxia or extrapyramidal signs. Both long and short term memory were preserved. In consultation with our psychiatrist, we applied the mini-mental state examination (MMSE), and the patient scored 28/30 (>23 being normal).[6] Orientation, attention, calculation and speech fluency were normal. The Geriatric Depression Scale score did not indicate depression.

The patient was diagnosed as a case of Benson's syndrome. In consultation with our psychiatrist, since the patient lacked any cognitive loss, no medicines were prescribed. The patient was advised a healthy diet, regular exercise, and recommended for follow-up every 6 months, with the freedom to contact us anytime he needed to.

Discussion

Given the history, clinical findings and supporting investigations, our patient fits into the profile of a case of Benson's syndrome. This is borne out by the fact of its presenile insidious onset, primarily with visual presentation, and cortical atrophy centered in the occipital cortex, while sparing memory and cognition: The last two more characteristic of AD. Yet, since both diseases are progressive in nature, there occurs considerable overlap. Simple tests of visual perception and copying can help to differentiate the two disorders. Because tasks with significant visual component (e.g., visual memory recall) may not be very valid in the presence of posterior cortical atrophy, those with minimal visual demands (e.g., auditory-verbal memory tasks, naming from verbal description) are therefore required for accurate cognitive assessment.[3] We used the MMSE which showed normal cognition and memory. If available, single photon emission computed tomography and fluorodeoxyglucose positron emission tomography have reportedly shown selective hypometabolism and hypoperfusion in the occipito-parietal region, worse on the right hemisphere, in cases of Benson's syndrome as compared to AD.[7] Cross-sectional voxel-based morphometry is reported

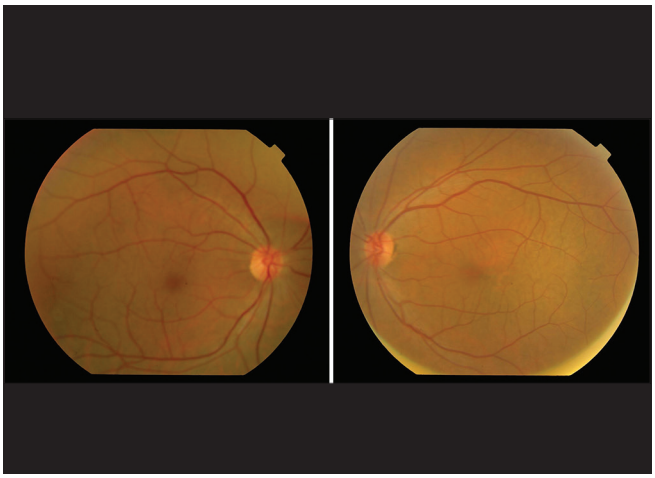

Figure 1: Fundus photographs show normal fundus, OU

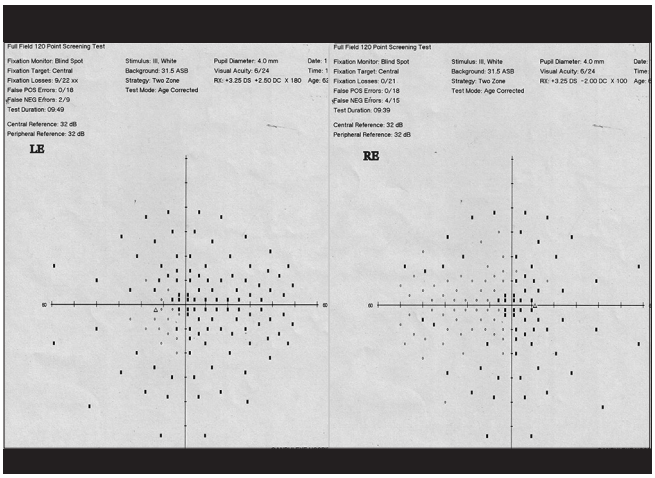

Figure 2: Full-field perimetry shows bilateral field defect, denser in the right fields, but also involving the periphery of the left fields in both the eyes

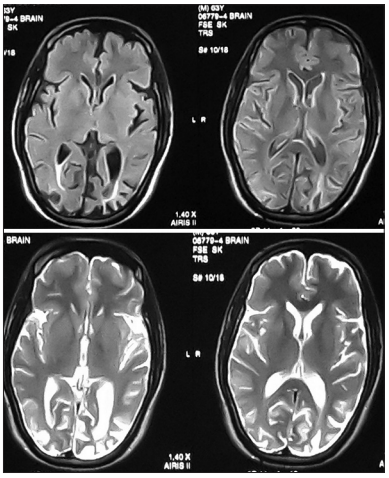

Figure 3: Magnetic resonance imaging brain shows prominent cortical sulci in bilateral occipital lobes with prominent occipital horns with mild periventricular hyperintensities on T2-weighted and fluid-attenuated inversion recovery images

to have revealed significant reductions in regions of the occipital and parietal lobes, and only later in the temporal lobe.[8] However, the neuroimaging findings of the two, merges over time. Also the neurofibrillary tangles and neuritic plaques characteristic of AD, have been found in the autopsy of Benson's syndrome patients, distributed mainly in the posterior cortical areas.[3] These features suggest that Benson's syndrome is an atypical variant of AD. Acetyl cholinesterase inhibitors, e.g., donepezil, rivastigmine and galantamine are frequently prescribed, given that AD is statistically the most likely underlying pathology, with limited single case reports suggesting some clinical benefit, most likely in patients with underlying Alzheimer's.[9] Anti-depressant medications may be given in patients with the persistent low mood. Cognitive rehabilitation including psychoeducation, compensatory strategies and cognitive exercises improve functioning in Benson's syndrome.[10] Since in our patient cognition was normal with no symptoms of depression, except for regular follow-up, no medical therapy was advised.

This case highlights the fact that on account of the overwhelming visual problems, an ophthalmologist may first confront such patients. Only with a high degree of suspicion will attention be drawn to such a possibility, and unnecessary vision enhancing surgery and pointless multiple change of glasses be avoided.

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